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RAREST AMONG THE RARE – PYLORIC ATRESIA – A CASE SERIES

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ABSTRACT

Gastric outlet obstruction in a neonate can occur due to several aetiologies. Here I am presenting 4 cases of the same. 1st is a case of a 4 days old male child who was brought with complaints of persistent non bilious vomiting since D1 of life. Examination revealed the presence of visible gastric peristalsis. X ray abdomen done showed the presence of a dilated stomach with absent distal gas shadow. USG abdomen done showed normal SMV SMA axis. Intraoperatively a type B pyloric atresia was noted for which a gastroduodenostomy was done. Post operative period was uneventful and the child was discharged on POD 8. 2nd is a case of a 12 days old male child who was admitted with complaints of respiratory distress and was intubated for the same .Child was extubated on day 10 and was started on oral feeds, following which the child had persistent vomiting which was non bilious . X ray abdomen taken showed a dilated stomach with paucity of distal gas shadow. Barium meal follow through done showed stasis of contrast in the stomach even in 6 hrs. Child was taken up for a laparotomy and was found to have a type A pyloric atresia. Gastrotomy was done and the membrane was excised. Post op child continued to have vomiting and underwent a repeat barium study which showed stasis in the stomach. Child was then taken up for a laparotomy and a gastrojejunostomy was done. Child was discharged and is on follow up. Among the entire spectrum of anomalies causing GOO, pyloric atresia is a very rare condition (1 in 100,000 newborns) Successful management is dependent on the early recognition of the entity and appropriate surgical intervention, optimum post-operative care and absence of associated anomalies.

INTRODUCTION

Pyloric atresia is an extremely rare condition which accounts for less than 1% of all atresias and diaphragms of the GI with an incidence of 1 in 1,00,000. It is said to occur because of an arrest in the development between 5th and 12thweek of gestation. The first case of congenital pyloric atresia (CPA) was reported as early as in 1749 by Calder but the first successful operation for the same was done only in 1940 by Touroff. Here we presenting a series of 4 cases of CPA

Case 1

1st born term male child was brought on the 4th day of life with complaints of non-bilious vomiting since the 2nd day of life. There was no history of abdominal distention noted by the mother. antenatal scans revealed there to be polyhydramnios in the mother. There was no history of gestational diabetes mellitus in the mother. The child had been delivered by caesarean section and had a birth weight of 2.7kg. The child had passed meconium on Day 1 of life and was started on feeds subsequently. On examination the child was irritable with a weak cry and reduced activity. Child was dehydrated. There were no skin lesions noted. On examination of the abdomen there was upper abdomen fullness noted with a visible gastric peristalsis. There was no abdominal wall erythema or oedema. Anal orifice was normal and external genitalia was also normal. Routine investigations showed elevated renal parameters which reverted to normal after the child was hydrated. An X ray abdomen was taken which showed a grossly dilated stomach with absent distal gas shadow. Ultrasound abdomen was done which showed a normal SMV SMA axis with no other anomalies (Fig 1). A preoperative diagnosis of a pyloric obstruction? atresia was made. Child was taken up for a laparotomy and intraoperatively was found to have a type B pyloric atresia where in the pylorus was one solid tube without a lumen(Fig 2) . We proceeded with resection of the pyloric segment followed by a gastroduodenostomy (Fig 3). Post- operative period was uneventful and the child was started on oral feeds on post - operative day 4. Child was discharged on POD 8

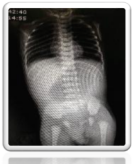

Fig 1

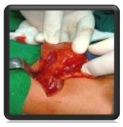

Fig 2

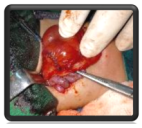

Fig 3

2nd case

12 days old ,term male child was referred with complaints of respiratory distress since day 5 of life. There was no history of vomiting in the child. Antenatal scans done showed no evidence of polyhydramnios and the mother had no co morbidities. The child had passed meconium on Day 1 of life and was on oral feeds which was subsequently discontinued in view of distress. The child on arrival was having significant respiratory distress and was intubated for the same. A nasogastric tube was inserted without a hitch. X ray chest and abdomen taken showed a pneumonic patch in the right upper lobe and a prominent stomach shadow with presence of minimal distal bowel gas. Child was extubated on day 10 and was started on oral feeds following which the child had persistent vomiting which was non - bilious. X ray abdomen taken showed a dilated stomach with paucity of distal gas shadow. Ultrasound abdomen was taken which did not show an altered axis. Barium meal follow through was done showed stasis of contrast in the stomach even in 6 hrs with a faint distal passage of contrast. A provisional diagnosis of pyloric atresia was made and the Child was taken up for a laparotomy and was found to have a type A pyloric atresia. Gastrotomy was done and the membrane was excised. Post operatively child continued to have increased non- bilious aspirate via the NG tube. Repeat X ray taken showed paucity of distal gas shadow with a dilated stomach. Child underwent a repeat barium study which showed stasis of contrast in the stomach. Child was then taken up for a Relaparotomy and a gastrojejunostomy was done. Child improved subsequently and repeat contrast study done showed entry of contrast into the distal bowel. Child was discharged and is on follow up.

Fig 4

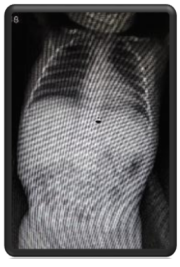

Fig 5

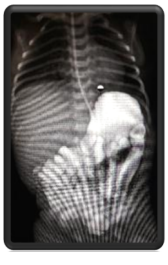

Fig 6

The above two cases and 2 other cases have been summarised in table 1

Table 1

	Case 1	Case 2	Case 3	Case 4
Age at presentation	4th day	12th day	5th day	2nd day
Sex	Male	Male	Male	Male
Polyhydramnios	Present	Absent	Absent	Present
Family history	Absent	Absent	Absent	Absent
Skin lesions	Absent	Absent	Absent	Absent
Dehydration	Present	Absent	Present	Present
Visible gastric peristalsis	Present	Absent	Absent	Present
Dyselectrolytemia	Absent	Absent	Absent	Present
Dilated stomach in X ray	Present	Present	Present	Present
Distal gas shadow	Absent	present	absent	Absent
Associated anomalies	Nil	Nil	Nil	ASD +
Type of atresia	Type B	Type A	Type A	Type B
Procedure	Gastroduodenostomy	Gastrotomy with membrane excision	Gastrotomy with membrane excision	Gastroduodenostomy
Post period	Uneventful	Relaparotomy with Gastrojejunostomy	Uneventful	Uneventful
Follow up	1 year	1 year 7 months	2 years	9 months

DISCUSSION

Gastric outlet obstruction in newborn period can be due to pyloric atresia, antral web or a hypertrophic pyloric stenosis with hypertrophic pyloric stenosis being the most common (1).

Congenital pyloric atresia (CPA) contributes to 1% of all atresias and diaphragms of the Gastrointestinal tract. 50% of these are associated with other anomalies, with epidermolysis bullosa (EB) particularly the junctional variant being the most common (1).The association between congenital pyloric atresia and epidermolysis bullosa is called Carmi syndrome or PA-EB syndrome (2) .

Familial occurrence of pyloric atresia has been reported (2)
Exact aetiology is still unknown and mucosal desquamation is said to play a role in its occurrence (2). Recently, mutations involving the genes which code for polypeptides, Integrin alpha 6 and Integrin 4 beta have been implicated in its occurrence (3). One hypothesis suggests that CPA occurs as a result of intrauterine complication of EB where the pyloric mucous membrane is affected leading to sloughing with subsequent scarring and fibrosis and obliteration of the pyloric canal (4)

Types of pyloric atresia are
A – membranous obstruction (most common)
B – longitudinal segmental atresia
C – Pyloric aplasia

Presence of polyhydramnios with a dilated stomach is a common feature in antenatal scans (3). In 1990, Meizner and Carine described the Snow Flake sign, an echogenic appearance of the amniotic fluid during the second trimester of pregnancy associated with epidermolysis bullosa (4)

There is no gender predominance noted in pyloric atresia. Prematurity is a common feature in this condition. Most common symptom is non- bilious vomiting and on examination there is presence of epigastric distension. Respiratory symptoms are common (3). Delayed diagnosis can result in gastric perforation as early as 12 hrs post- delivery (3).

Plain X ray abdomen and chest will show the presence of a dilated stomach and absent distal gas shadow. Contrast study will show complete obstruction at the pyloric region. Presence of radiological signs namely – 1) The single gas bubble
2) Absence of a beak sign
3) Presence of a pyloric dimple sign on contrast studies

Is diagnostic of pyloric atresia.
It is to be remembered that single gas bubble sign by itself is not pathognomonic of pyloric atresia. It is indicative of a gastric outlet obstruction(4)

USG abdomen can be done which can assess the Superior mesenteric vein and artery axis and can also help by demonstrating the absence of normal pyloric muscle and canal.

Initial management involves initial fluid resuscitation and insertion of a nasogastric tube to decompress the stomach. Electrolyte imbalances have to be corrected. Central venous catheter insertion is recommended as the child may require total parenteral nutrition and prolonged medical treatment especially if associated epidermolysis bullosa is present (2)

Depending on the type of pyloric atresia, the operative procedures vary. Membranous atresias are best tackled by excision of the membrane with pyloroplasty according to either Heineke – Mikulicz or Finney (1). Transgastric excision of pyloric membrane without pyloroplasty has also been reported (1).

Type B and C atresias are best managed by excision of the segment and end to end gastroduodenostomy in the former and only an end to end gastroduodenostomy in the latter (1).

Gastrojejunostomy is not usually recommended due to an increased risk of marginal ulcers and blind loop syndrome (1). However it may be carried out as a life - saving procedure in redo surgeries (4).

CONCLUSION

Presence of supra ampullary obstruction should be suspected in a newborn with persistent non - bilious vomiting and upper abdominal distension. Even though an association with epidermolysis bullosa is an unique feature of CPA, it can occur even in its absence. Successful management is dependent on the early recognition of the entity and appropriate surgical intervention, optimum post-operative care and absence of associated anomalies

ABBREVIATIONS

CPA - Congenital pyloric atresia
EB – Epidermolysis bullosa

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